

presses of witch-hazel or, preferably, eucupin, 1-100, are kept on the operative area. He may rise to micturate, and if he has difficulty a sitz bath will help. On the second day he is given full diet, and the third night a laxative is given and usually he may go home on the 4th day. We usually begin liquid paraffin every night following the operation, and this should be continued for a week or so. For local tenderness, as a lubricant I have been using with very satisfactory results an ointment of eucupin, 1 per cent, in eucerin.

Note.—Since preparing the above paper, following methods advocated by Manheim and Marks,² I have been using for local anæsthesia novocaine, 1 per cent, with eucupin, 1-1000, and find it even more efficient than the procedure described in the paper.

REFERENCES

1. MORGAN, C. N.: Oil-soluble anæsthetics in rectal surgery, *Brit. M. J.*, 1935, 2: 938.
2. MANHEIM, S. D. AND MARKS, M. M.: Eucupin (isoamylhydrocupreine) as local anæsthetic in proctologic surgery and in the treatment of pruritus ani, *Am. J. Surg.*, 1938, 39: 86.

KRAUROSIS, LEUKOPLAKIA AND PRURITUS VULVÆ: TREATMENT BY RESECTION OF THE SENSORY NERVES OF THE PERINEUM*

By B. USHER, M.D. AND A. D. CAMPBELL, M.D.

Montreal

THIS article is a report regarding resection of the sensory nerves of perineum in cases of kraurosis, leukoplakia and pruritus vulvæ with lichenification.

There is much confusion in the literature concerning the definition of the terms. Montgomery and his co-workers¹ have given us an exhaustive clinical and pathological study of the whole subject. They believe that the three conditions represent distinct entities and that one may occur independently of either of the other two. It is true, too, that two or all three of the conditions may be present clinically or pathologically, or both, and may merge with one another, especially as all three are likely to appear in the late decades of life. This merging of the three conditions accounts for the confusion in terminology.

Darier's definition of kraurosis cannot be improved upon: "The term kraurosis should be confined to a sclerosing progressive atrophy of the mucocutaneous teguments of the vulva, leading gradually to stenosis of the vaginal orifice, the disappearance of the labia minora, the frenulum and the clitoris and flattening of the labia majora. The mucosa of the parts involved is always shining, smooth and dry. The colour is white, waxy yellow, red or spotted. Complication by leukoplakia is frequent."²

The histopathological changes in kraurosis are essentially those of an atrophic process. There is relative and often absolute hyperkeratosis, with little change in the stratum granulosum. There are marked atrophy of the prickle-cell layer and liquefaction necrosis of the basal-cell layer; an area of œdema just beneath the epidermis with œdema and homogenization of the connective-tissue fibres; obliteration of the rete ridges as a result of the œdema; homogenization of the connective-tissue in the upper portion of the cutis; a perivascular infiltrate in the mid-cutis composed of lymphocytes and later fixed connective-tissue cells; there is more or less atrophy of the epidermal appendages. In the later stages a varying degree of parakeratosis is present and sclerosis of the deeper vessels may occur.

Leukoplakia of the vulva presents features comparable to leukoplakia of the mouth. Clinically, it occurs as single or multiple discrete plaques involving the inner surfaces of both labia, the perineum, and the clitoris, or the vaginal mucosa. Leukoplakia is prone to occur near the clitoris and on the perineum and inner surfaces of the labia minora. It is in these sites that the pruritus is most intense. It is greyish to greyish-blue in colour, with a thickening of the skin or mucous membranes with no scaling. As the leukoplakic process progresses the epidermis becomes markedly thickened and fissures or ulcerations may appear.

* Read before the Sixty-eighth Annual Meeting of the Canadian Medical Association, Section of Dermatology, Ottawa, June 24, 1937.

From the Departments of Dermatology and Gynecology, Montreal General Hospital.

The histopathological change in leukoplakia is essentially a hypertrophic process. There is early hyperkeratosis, an increase in the stratum granulosum, acanthosis, and relatively little change in the cutis. Soon one sees a perivascular as well as a diffuse infiltrate, chiefly lymphocytic, in upper portions of the cutis, with varying degrees of liquefaction-necrosis of the basal cell layer. Dependent on the degree of infiltration in the cutis, a varying degree of destruction of the elastic tissue occurs, and different degrees of fibrosis with obliteration of the lumina of the superficial vessels may be seen.

Persistent and chronic pruritus vulvæ results in lichenification, with most frequently involvement of the outer aspects of the labia majora, about the pubic hairs, the inner aspects of the thighs, and also the anus. At times the inner surfaces of both labia, the clitoris and the perineum may be involved. Clinically, one finds discrete or diffuse plaques of thickening of the skin, with accentuation of the normal markings. When moisture is present, the lesion assumes a whitish soggy appearance.

Histopathologically, there are hyperkeratosis alternating with parakeratosis, acanthosis with prolongation of the rete pegs, elongation of the papillary bodies and a mild perivascular lymphocytic infiltrate in the upper part of the cutis, with some dilatation of the superficial vessels. No liquefaction necrosis of the basal cell layer is present. There are no appreciable changes in either connective or elastic tissue. Pruritus is the predominant symptom in all these conditions.

Little is definitely known as to the etiology of the various lesions grouped under pruritus vulvæ. It is worthy of note, however, that the general distribution on the affected area corresponds geographically with the so-called sex skin in the primate. It is possible that certain persons possess to a degree an area about the external genitalia which responds to the œstrin tide as does that of the monkey. At or after the menopause such tissue undergoing certain structural changes possibly gives rise to the various clinical entities herein described. On the other hand, only a small percentage of those suffering from pruritus are relieved by the administration of œstrogenic substances at present available. Such an observation, however, does not wholly disprove the general thesis of the relationship of regressional changes in the vulva to alterations

in the physiological activity of the glands of internal secretion.

TREATMENT

In the past treatment has consisted in the local application of ointments and lotions, while x-ray and, more recently, the glandular extracts have been extensively employed. Since August, 1934, resection of the sensory nerves of the perineum has been performed on 8 patients in whom pruritus proved refractory to non-surgical measures. The technique described by Learmonth, Montgomery and Counsellor,³ with but minor modifications, was adopted.

Sensory nerve resection has been practised sporadically during the past eighty years or more. It is interesting to observe that Sir J. Y. Simpson⁴ employed a technique for nerve resection described by Dr. Burns, of Glasgow, in his "Principles of Midwifery". Since the perineal area is not supplied by a single nerve, but rather by branches of five different nerves, namely, the ilio-inguinal and genito-crural nerves, dorsal nerve of clitoris, superficial perineal nerve, pudendal branch of small sciatic nerve, and inferior hæmorrhoidal nerve, identification of these obviously offers a major anatomical problem.

While it is true that regressional changes do occasionally follow perineal sensory nerve resection, one must not lose sight of the fact that carcinoma in this region commonly follows leukoplakia, with pruritus the only symptom and excoriation from trauma the only visible lesion. On the other hand if the biopsies taken at the time of the nerve resection do not reveal malignancy this procedure will relieve symptoms and does not preclude vulvectomy (an operative measure of a definitely radical nature) for carcinoma of the vulva if and when it is detected.

CASE 1

Mrs. A.K., aged 35, presented herself at the dermatological out-patient department of the Montreal General Hospital with pruritus of the vulva of six years' duration. Examination revealed a single patch of lichenification on the mons veneris. In 1935 an artificial irradiation menopause had been produced with no effect on the pruritus. Examination otherwise revealed normal findings. Biopsy of the affected part showed the typical histopathological feature of lichenification. Section of the perineal and pudendal nerves was performed in June, 1936. There was immediate relief from the pruritus and disappearance of the local lesion. To date, twelve months later, there has been no recurrence.

CASE 2

Mrs. J.W., aged 63, whose menopause had occurred at the age of 45, was under treatment in the dermato-

logical out-patient department for four years with an erythematous-squamous eczema of the crural region, the lower abdomen and labia majora. There was a great deal of thickening of the parts and the pruritus was extreme. Local therapy, irradiation and various diets, including a low carbohydrate diet because of a blood sugar reading of 0.131 mg. per cent, were of little benefit. Physical examination otherwise revealed normal findings. No biopsy was performed. Section of the sensory nerves of the perineum was performed in September, 1934. The patient remained well for the ensuing twelve months, at which time she disappeared from observation.

CASE 3

Mrs. E.C., aged 47, was under observation for 15 years for an intractable pruritus of the vulva which was so severe as to cause considerable loss of sleep. Her menopause occurred three years subsequent to the onset of the pruritus. Examination of the genitalia in September, 1934, revealed an outlet small and shrunken, with disappearance of the labia minora. The vestibule was pale and flat-surfaced, and this pale atrophic surface extended along the inner sides of both labia majora to include the clitoris. There was a small patch of leukoplakia on the inner aspects of the labia minora. Physical examination, including blood sugar, was otherwise normal. Section of a piece of affected tissue showed the histopathological features of kraurosis vulvæ (Fig. 1). Section of the

prior to observation the pruritus had been so severe as to require sedatives for its relief. Physical examination, including blood sugar estimation, revealed normal findings. Nerve resection was performed on March 25, 1936, with immediate relief from the pruritus. She reported by letter, dated May 6, 1937, that marked relief from the pruritus persisted in spite of the presence of some erythema in the groins.

CASE 5

Mrs. C.W., aged 49 (case of Dr. R. Powers), suffered from pruritus vulvæ of eleven years' duration which caused loss of sleep and extreme irritability. Menstruation was still normal and physical examination revealed no pathological change except lichenification of the outer aspects of the labia majora. Nerve resection was performed by Dr. Powers in October, 1935, and to date there has been complete relief from the pruritus with disappearance of the lichenification. No biopsy was performed.

CASE 6

Mrs. W.B., aged 53, complained of pruritus vulvæ of ten years' standing. Examination revealed the picture of kraurosis vulvæ and secondary lichenification, with physical findings otherwise normal. Attempts at resection of the nerves were on two occasions only partially successful. There was only partial relief from the pruritus which still persisted to a somewhat lesser degree at the date of writing. His-



Fig. 1

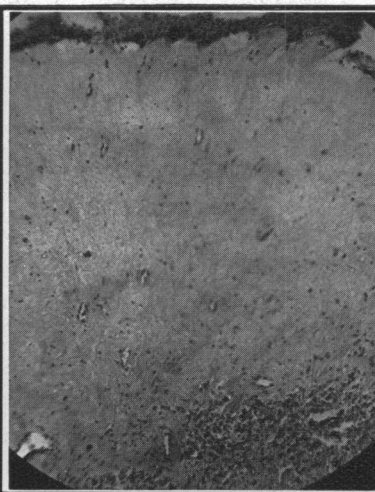


Fig. 2

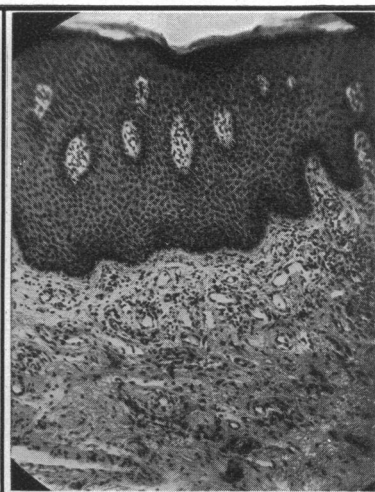


Fig. 3

Fig. 1 (Case 3).—Typical histological picture of a case of kraurosis vulvæ, showing hyperkeratosis, atrophy of prickle-cell layer and oedema and homogenization of the connective-tissue. Fig. 2 (Case 6).—Histological picture of a case of kraurosis vulvæ. Note the collection of lymphocytes in the lower right corner. Fig. 3 (Case 8).—Histological picture of neuro-dermatitis of the vulvæ showing acanthosis, papillomatosis, dilated capillaries and lymphocytic infiltrate.

perineal nerves was performed in September, 1932. With the exception of mild pruritus in April, 1937, due to a vaginitis, there has to date been complete relief from subjective symptoms. There had been no change in the appearance of the kraurosis, but the leukoplakia had disappeared.

CASE 4

Mrs. B., aged 48 (patient of Dr. J. F. Burgess), came under observation in March, 1936, for pruritus vulvæ et ani. Examination showed the presence of erythematous lichenified eczema of the groins and labia majora and in the region of the mons veneris. The anal pruritus dated back to the days of her youth, while the pruritus vulvæ first appeared in 1929. Local applications, including x-ray therapy, had been of only temporary benefit, and for the four months

tological examination of tissue showed in two different sections both kraurosis and lichenification (Fig. 2). Lack of success in this case could be attributed to failure to excise all the nerves.

CASE 7

Miss Vera S., aged 29, was seen for a pruritus vulvæ of two years' duration and pruritus ani of ten years' duration. Nerve section was performed in January, 1937. There was complete relief of her symptoms at the time of discharge from the hospital. No follow-up has been possible in this patient.

CASE 8

Mrs. H.A., aged 37, complained of pruritus vulvæ of nine months' duration. There was no response to progynon and ointments applied locally. Physical ex-

amination revealed lichenification of the outer aspects of the labia majora with nothing of significance otherwise. Histopathological examination showed lichenification (Fig. 3). Section of the sensory nerves was done in November, 1936, with complete relief from pruritus and no recurrence at the time of writing.

Resection of perineal and pudendal nerves of the perineum has been performed on eight patients, six with lichenification and two with kraurosis vulvæ. These patients have been observed for periods ranging from five months to two and one-half years. In cases 1, 2, 4, 5 and 8, complete relief was obtained for periods varying from five months to two and one-half years. In case 3 there was complete relief for two and one-half years, when vaginitis and leucorrhœal discharge resulted in a slight recurrence which rapidly subsided under appropriate treatment. It was also noted that the leukoplakia patch had also disappeared. In case 6 the nerves could not be completely excised. In case 7 relief from symptoms was obtained, but the period of post-operative observation is as yet too short to determine the outcome. Thus in all our cases where resection of the nerves was complete the results were most gratifying. It is true, however, that the majority of these patients have not been followed sufficiently long to determine the permanency of relief. It must be pointed out that in the majority of cases operated upon local applications, x-ray therapy, and endocrine therapy had been previously employed without success.

We wish to point out that results depend on the care taken to exclude those patients with conditions which contraindicated operation: (1)

those with local or general disturbances which might produce vulvar pruritus; (2) those showing malignant changes or severe radio-dermatitis; (3) markedly obese patients. In one such case where it was attempted the nerves could not be completely removed; (4) psychoneurosis.

CONCLUSION

Kraurosis vulvæ, leukoplakia and pruritus with lichenification are distinguishable from one another on clinical and pathological grounds. In many cases all three may merge.

Resection of the sensory nerves of the perineum is contraindicated in the presence of malignant change or actinodermatitis. Resection is also contraindicated in the presence of psychoneurosis.

Resection is definitely the treatment of choice in cases refractory to other methods of treatment. It affords immediate and permanent relief in a large percentage of selected cases. It is therefore felt that such a procedure, by relieving the irritation and constant trauma from scratching, possibly prevents subsequent carcinomatous change developing in the area involved. It is worthy of note that in one patient with kraurosis vulvæ and secondary leukoplakia, regression of the leukoplakia resulted.

REFERENCES

1. MONTGOMERY, H., COUNSELLER, V. S. AND CRAIG, W. McK.: Kraurosis leukoplakia and pruritus vulvæ, *Arch. Dermatol. & Syphilol.*, 1934, 30: 80.
2. Quoted by Montgomery et al.: *Arch. Dermatol. & Syphilol.*, 1934, 30: 84.
3. LEARMOWTH, J. R., MONTGOMERY, H. AND COUNSELLER, V. S.: Resection of sensory nerves of perineum in certain irritative conditions of external genitalia, *Arch. Surg.*, 1933, 26: 50.
4. SIMPSON, J. Y.: Quoted by Learmowth et al.

XANTHOMATOSIS OSSIUM*

BY GORDON S. FRENCH, M.D.

Owen Sound, Ont.

MANY of the conditions of generalized bone disease present no difficulty of diagnosis and respond to a particular plan of treatment. This is particularly true of the generalized bone disease known as osteitis fibrosa cystica, also spoken of as Recklinghausen's disease or hyperparathyroidism. The diagnosis is based on the finding of cystic lesions in the bones by x-ray examination, together with the associated blood

picture of hypercalcæmia and hypophosphatæmia, plus increased excretion of calcium in the urine leading to a negative calcium balance. The removal of a parathyroid tumour is followed by a rather remarkable recovery.

If, on the other hand, the cystic bone lesions are accompanied by exophthalmos, diabetes insipidus, and xanthomatosis cutis the condition of Schüller-Christian disease is readily recognized, and the finding of "foam cells" by biopsy substantiates the diagnosis. However, patients with generalized bone disease are met with from

* Read at a meeting of the Baltimore City Medical Society (Radiological Section), Baltimore, Md., October 15, 1935.